CASE REPORT

AGGRESSIVE GIANT CELL TUMOR OF THE ANTERIOR RIB WITH THORACIC EXTENTION: A CASE REPORT AND REVIEW OF LITERATURE

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ABSTRACT

A giant cell tumor is a benign bone tumor, accounting for 4% to 9.5% of all bone tumors and 20% of all benign bone tumors. Ribs are rare site for giant cell tumor and only few cases have been reported in the literature with most of them involving the posterior aspect. We report a case of a large giant cell tumor in a 12 year old boy involving left anterior 4th rib with intra and extrathoracic involvement.

Key words: Giant Cell Tumor, Benign Tumor, Anterior Rib, Aggressive, Thoracic

Introduction

A giant cell tumor is a benign bone tumor, accounting for 4% to 9.5% of all bone tumors and 20% of all benign bone tumors. It occurs mainly in the proximal tibia, humerus, distal radius bone and the pelvic bone.[1] It is rarely observed in sites such as ribs and temporal bone, though local recurrence rate is up to 5% to 25%.[2] These are usually found posteriorly (epiphysis of head and tubercle), and their presentation anteriorly is very rare.[3] Giant cell tumors are often associated with local invasion, recurrence and metastases, and neither age nor gender can predict the metastases.[4] We report a case of a large giant cell tumor in a 12 year old boy involving left 4th rib with intra and extrathoracic involvement.

Case Report

A 12-year-old male presented with a four-month history of a progressively growing mass in the left anterior chest wall not associated with pain. There was no significant past history but family history of tuberculosis was positive. Initially, patient took antibiotic treatment multiple times but the mass progressively increased in size. On local examination, there was a huge swelling on anterior aspect of left upper chest wall. It was firm to hard in consistency, fixed to the underlying structures and chest wall. The overlying skin was normal. The patient was vitally stable, however, had decreased air entry in the left upper chest. No enlarged local lymphadenopathy was present. All other systemic, hematological and biochemical investigations were within normal limits.

Patient was sent for the radiological workup and CXR showed destruction, erosion and expansion of the left 4th rib with a large soft tissue component with intra and extrathoracic extension. Initially, a differential diagnosis of ewing's sarcoma, giant cell tumor with malignant transformation and osteosarcoma was made.

CT Scan chest with contrast was done to evaluate the mass lesion. CT showed a large destructive bubbly, expansile mass lesion arising from the angle and anterior part of the left 4th rib causing significant

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destruction and erosion. The lesion was infiltrating the left anterolateral chest wall with extrathoracic component bulging into subcutaneous soft tissues. It has an intrathoracic, heterogenously enhancing, soft tissue component with a large central necrotic area causing significant atelectasis in left lung predominantly lower lobe. (Fig. 1) Medially, the lesion was abutting the paravertebral region, left atrium, left

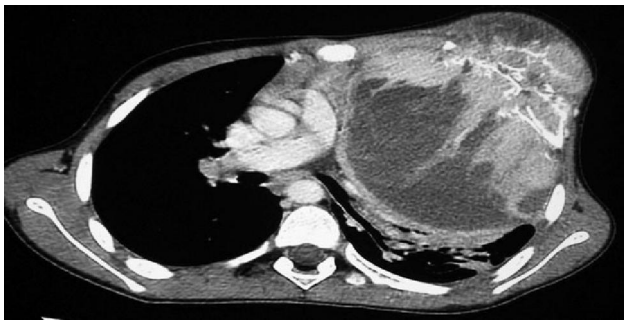

Figure 1: Axial section of CT Scan Chest with contrast showing a large soft tissue lesion with intra and extrathoracic components arising from the left 4th anterior rib with its destruction and complete erosion. It is causing compression collapse of the left lung. The lesion is showing central areas of necrosis. Biopsy proven Giant cell tumor of left anterior fourth rib.

ventricle and left upper pulmonary vein without encasement or infiltration. It was causing displacement of the mediastinum to the right side. Inferiorly, it was abutting the left dome of diaphragm and posteriorly, left lateral chest wall. (Fig. 2) It was measuring 16.5 x 12.5 x 11.0 cm in anteroposterior, craniocaudal and transverse dimensions. Mild pleural effusion was also present, however, no enlarged mediastinal lymphadenopathy was seen. A small nodule was present in the laterobasal segment of right lung lower lobe suggestive of metastasis.

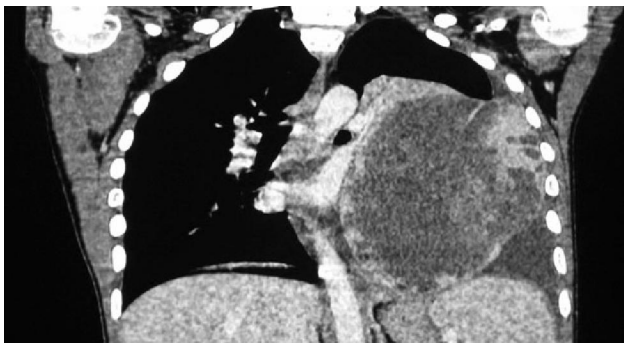

Figure 2: Coronal section of CT Scan Chest with contrast showing a large tumor mass lesion causing shifting of the mediastinum towards right side.

Further workup with bone scintigraphy was performed using Technetium-99m Methyl Disphosphonate (Tc^{99m} MDP), that showed intensely increased tracer uptake in left side of the chest wall representing site of primary tumor. Rest of the scan was negative for distant spread of disease.

Biopsy of the lesion was done and histopathology showed scattered clusters of multinucleated giant cells with bony spicules and dilated blood cells. No increased mitotic activity, necrosis or nuclear pleomorphism was noted. A final diagnosis of Giant cell tumor was made.

Discussion

The incidence of Giant cell tumor among chest wall tumors is about 1-2%.[5] The incidence is lower in the sternum than in the ribs. It is rare in immature skeletons and maximally presents in the third and fourth decades. There is a mild female preponderance. Few cases involving giant cell tumors occupying the thoracic cavity have been reported in the literature.[6,7] Many large series have reported an incidence of around 1% in the ribs; after reviewing 15 cases, Gupta and Mittal showed that most of these involved the posterior aspect of the rib.[3] There are only few published cases of giant cell tumors arising from anterior arc of the rib.[8]

Giant cell tumors are aggressive tumors and present with the signs and symptoms of pain, swelling, and limitation of motion about a joint. Hutter et al. report that patients experienced symptoms for an average of 10 months prior to first treatment of benign Giant Cell Tumor (GCT).[9] However, our patient noted a progressively growing mass over a period of 3 to 4 months, and the occasional pain began near his time of presentation. This probably contributed to the extremely large tumor size found at the time of presentation. While multiple cases of GCT originating from the rib have been reported, this case is unique in its presentation with both intra and extra thoracic soft tissue component.

Variants of GCTs include chondroblastoma, chondromyxoid fibroma, aneurysmal bone cyst, and “brown”

tumor of hyperparathyroidism.[10] When differentiating GCTs of rib from simple bone cyst Oschner described that the latter are more likely to be formed in the anterior part of the ribs, whereas GCT are mostly located posteriorly in the epiphysis of bone (i.e., the head and tubercle of ribs).[11] Only 3% of GCTs develops in the immature skeletons which distinguishes these patients from those with aneurysmal bone cysts, in whom the tumor maximally occurs prior to epiphyseal fusion.

Plain radiographs of these tumors show eccentric osteolytic lesions accompanied by cortical thinning and expansion. CT allows evaluation of the extent of the tumor and its relationship to surrounding structures. CT improves detection of cortical thinning, pathologic fracture, periosteal reaction, and degree of osseous expansile remodeling compared with radiograph.[12,13] The solid portions of GCT demonstrate attenuation similar to that of muscle. Soft-tissue extension is common at CT and MR imaging and was seen at CT in 33% - 44% of cases in studies by Levine et al[13] and Hudson et al.[12] As with other musculoskeletal neoplasms, MR imaging is superior to CT in delineating soft-tissue tumor extent because of its improved contrast resolution. GCT shows low intensity on T1W and heterogeneous high intensity on T2W images. Therefore intramedullary tumor is best seen on T1W, while its extraosseous portion is best appreciated on T2W images.[14,15] GCT produces increased uptake of technitium-99m radio-pharmaceuticals. The pattern of increased uptake may be diffuse (40%) or peripheral with little central activity (60%).[16] The role of angiography today, in patients with GCT, is limited to a study of regional vascular anatomy and perhaps, transcatheter arterial embolisation in instances of unresectable neoplasms.[17]

Most postoperative metastases occur in the lungs, with a reported incidence between 1% and 9%[18,19] followed by the mediastinum and aortic lymph node[20] skin[21] and the breasts.[22] The mortality associated with lung metastases of giant cell tumors is 23%.[19] Considering that giant cell tumors are borderline tumors, most reports suggest that they are not responsive to chemotherapy even after the appearance of the lung metastases. Our patient presented with a metastatic pulmonary nodule in the laterobasal segment of right lung lower lobe preoperatively.

Current methods available to treat GCT include curettage, bone graft and complete surgical resection of the affected segment of bone.[23] Excision is desirable as 10% of GCTs in ribs undergo malignant transformation,[10] while radiation therapy is not recommended as most of malignant transformations are related to previous radiation therapy.[5] Thus en bloc excision is an appropriate treatment, and disease-free survival is directly proportional to negative resection margin. Hutter et al. report that most recurrences (81%) appear in less than 2 years, and almost all have been manifested by 4 years. Thus at least 5 years of close follow up are recommended. However, it has been reported that the course of a benign giant cell tumor undergoing malignant transformation may take longer than 5 years.[9]

Conclusion

Ribs are a rare site for giant cell tumor and only few cases of GCT involving the ribs have been reported in the literature with most of them involving the posterior aspect. Due to its rarity, GCT arising from the chest wall is difficult to diagnose, especially when the tumor is located in the anterior arc of the ribs.

CONSENT

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

CONFLICT OF INTERESTS

The authors declare that they have no conflict of interests.

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